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Leukaemia Section

Short Communication

t(X;8)(q24;q24)

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Abstract

Review on t(X;8)(q24;q24), with data on clinics

Keywords

Chromosome X; chromosome 8; Blastic plasmacytoid dendritic cell neoplasm

Clinics and pathology

Disease

Blastic plasmacytoid dendritic cell neoplasm (BPDCN)

Note

BPDCN has been known with various names, including agranular CD4+ natural killer (NK) leukemia, CD4+/CD56+ hematodermic neoplasm, and blastic NK lymphoma. BPDCN malignant cells are derived from the precursors of plasmacytoid dendritic cells. It most commonly involves the skin. BPDCN is an aggressive neoplasm. BPDCN is often associated with a complex karyotype (review in Meloni-Ehrig 2017).

Epidemiology

In a series of 41 patients with BPDCN, five had a MYC rearrangement confirmed by FISH: one had a t(X;8)(q24;q24), one had a t(3;8)(p25;q24), two had a t(6;8)(p21;q24) MYC/SUPT3H, and one had a t(8;14)(q24.1;q32) (Boddu et al., 2018).

Clinics

The patient with a t(X;8)(q24;q24) was a 3 year-old girl. She was alive and well 20 months+ after diagnosis.

Cytogenetics

The karyotype was complex, with del(6q), del(9q), and other abnormalities.

Genes involved and proteins

Note The partner gene of MYC is unknown.

MYC

Location 8q24.21

DNA/RNA

MYC is composed of three exons spanning over 4 kb.

Protein

MYC is expressed in almost all proliferating cells. It is located predominantly in the nucleus. MYC is a transcriptional regulator, capable to induce or repress the expression of thousands genes. MYC is deregulated in cancer by several different mechanisms: chromosomal translocations, amplifications, point mutations, epigenetic reprogramming, enhanced translation and increased protein stability (review in Mohamed, 2017).

References

Boddu PC, Wang SA, Pemmaraju N, et al. 8q24/MYC rearrangement is a recurrent cytogenetic abnormality in blastic plasmacytoid dendritic cell neoplasms. Leuk Res. 2018 Mar;66:73-78

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